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LETTER TO THE EDITOR

Recurrent lupus mesenteric vasculitis leading to gastrointestinal perforation and sepsis

To the Editor,

Systemic lupus erythematosus (SLE) is an autoimmune disease affecting multiple systems. Gastrointestinal (GI) manifestations are not included in the diagnostic criteria of SLE, but frequently affect SLE patients. There are various GI manifestations, including anorexia, nausea, and vomiting. The etiologies vary, such as lupus mesenteric vasculitis (LMV) and serositis secondary to lupus itself, side effects of SLE medication, or coincidence of GI diseases like appendicitis, pancreatitis, or acute viral or bacterial enterocolitis. Diagnosing the cause of GI symptoms in patients with SLE is a challenge for clinicians. However, LMV should be considered if all other etiologies have been excluded. Here is a case of a patient with SLE presenting as recurrent abdominal pain and with poor prognosis.

A 15-year-old girl had a history of SLE with class IV lupus nephritis and lupus nephritis-related secondary hypertension. On the first admission, she had abdominal pain for 2 weeks with fever, tarry stool, and vomiting for 4–5 days. Physical examination showed diffuse tenderness over the abdominal wall, without rebound tenderness or muscle guarding. Abdominal computed tomography (CT) revealed segmental mural thickening of the proximal jejunum. Acute segmental obstructive enteritis (ASOE), mesenteric vasculitis related, was highly suspected. As an SLE flare-up was favored, intravenous methylprednisolone (500 mg/d) was administered for 3 days. She was discharged in a stable condition, with oral prednisolone (1 mg/kg/d) as a maintenance treatment. However, she was re-admitted after 3 days with similar conditions, which were resolved with intravenous steroids.

Unfortunately, the patient had abdominal pain with vomiting, and watery diarrhea with blood-tinged stool 4 months later. Laboratory examination revealed a white cell count of 8180/uL (segment/lymphocyte: 97%/1%), platelet 73,000/mm^3, C-reactive protein 3.2 mg/dL, amylase/lipase 65/206 U/L, and SGOT/SGPT 53/45 U/L. Intravenous methylprednisolone (500 mg/d) was administered for 3 days, but the abdominal pain recurred. Abdominal X-ray of the kidneys, ureter, and bladder (KUB) demonstrated small intestine obstruction with proximal dilatation. Emergency abdominal CT (Fig. 1) revealed free air and fluid collection in the peritoneal cavity. Hollow organ perforation was highly suspected. There was also segmental wall thickening of the small bowel loop, as well as splenic infarction.

A pediatric surgeon immediately performed an exploratory laparotomy. Jejunal perforation was noted, so segmental resection of the jejunum with end-to-end anastomosis was performed. Histopathology report of the resected jejunum showed a picture of vasculitis of the small arteries, with fibrinoid necrosis of the vessel wall. Ulcer with perforation and acute inflammatory exudate in the serosal layer were observed. A broad spectrum antibiotic was administered due to the suspicion of sepsis. An antifungal agent was also administered because an ascites culture showed *Candida albicans*.

However, the abdominal pain recurred after the operation. Abdominal CT revealed focal abscess collection with peritonitis over the left abdomen. Surgical intervention was scheduled, but the patient had septic shock with consciousness change the next morning. Her consciousness remained confused even after drainage of the abscess. Brain CT revealed symmetric calcification over the bilateral basal ganglion, probably chronic SLE-related rather than acute change. The patient was intubated due to declining respiratory condition, but her condition worsened with multiple organ failure until she finally died.

A case report from Taiwan in 2011 involved a patient with severe, recurrent lupus enteritis and severe hypocomplementemia as the initial and only presentation of SLE. The patient died of sepsis [1]. This patient did not suffer from bowel perforation, but had total intestinal obstruction with obstructive jaundice resulting from obstruction of the ampulla of Vater, all of which were suspected to be secondary to severe duodenal wall swelling

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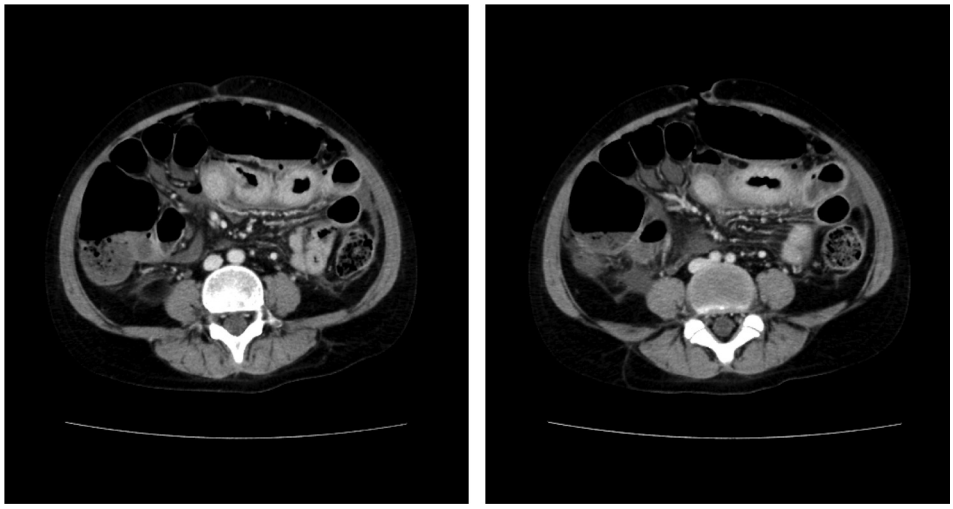

Figure 1. Abdominal computed tomography of the patient, showing free air and fluid collection in the peritoneal cavity, and hollow organ perforation was suspected.

and edema. This case and the present case report both had recurrent lupus enteritis that caused the poor prognosis. It remains unclear as to what risk factor of mesenteric vasculitis was related to poor prognosis.

There are several reports on recurrent lupus mesenteric enteritis. Kwok et al. [2] mentioned that SLE patients with pre-existing antiphospholipid antibody syndrome have a tendency for more frequent recurrences. Kim et al. [3] noted that the cumulative dose of prednisolone and the duration of prednisolone treatment are significantly higher in patients with nonrecurrent lupus enteritis than in those with recurrent lupus enteritis. The wall thickness of the most thickened bowel in patients with recurrent lupus enteritis is greater than that in patients with nonrecurrent lupus enteritis, although the difference is not significant [3]. Lian et al. [4] have found that patients treated with cyclophosphamide have less recurrent GI diseases.

The present case report had recurrent LMV for three times within half a year, despite antiphospholipid antibody titers within normal limits. She received steroid control for 6 years and intermittent intravenous cyclophosphamide treatment nine times, but she had poor medication compliance with only intermittent outpatient department visits. Thus, despite immunosuppressants and normal antiphospholipid antibody titers, poor drug adherence with lower cumulative oral prednisolone, and shorter treatment duration may have led to recurrent LMV. The report from Kim et al. [3] has 16 patients, including seven with recurrent LMV, but without any mortality. More clinical experience is needed to investigate risk factors of recurrent LMV and gastrointestinal perforation.

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